A CASE OF MOLLUSCUM FIBROSUM

(VON RECKLINGHAUSEN'S DISEASE).

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The case described below illustrates the usual features of molluscum fibrosum, and little is to be added to the original description of the disease published by von Recklinghausen in 1882. Several cases have been described and investigated since that date, and have all corroborated his discovery that the multiple growths occurring in the skin are of the nature of fibromata growing from the connective tissue of the terminal filaments of the cutaneous nerves. The standard work on the subject in this country is the exhaustive monograph by Alexis Thomson,[1] *On Neuroma and Neurofibromatosis*, published in 1900. Therein will be found a full description of the two cases recorded by von Recklinghausen, a complete account of symptoms, diagnosis, prognosis, etc., and a list of all the recorded cases of neurofibromatosis up to that date. Rolleston and Macnaughton[2] in 1912, in their paper on hereditary and familial types of the disease, give a further list of familial and hereditary cases published after 1900.

The patient here described is a man aged 60, a potter's slip-maker by occupation, who confesses to no previous illnesses, but who for the last few years has suffered more or less from chronic bronchitis. He is somewhat poorly nourished, but active both physically and mentally. He exhibits none of the mental dullness often associated with cases of von Recklinghausen's disease. There is no history of bilious attacks which have been described by some observers, nor can I find any evidence of pulmonary tuberculosis.

History.

He states that he first noticed small lumps on the skin of the chest about thirty years ago. These lumps appeared painlessly, and steadily increased in size, while fresh growths continued to appear. Some appeared to collapse after a time, but none of them have ever completely disappeared.

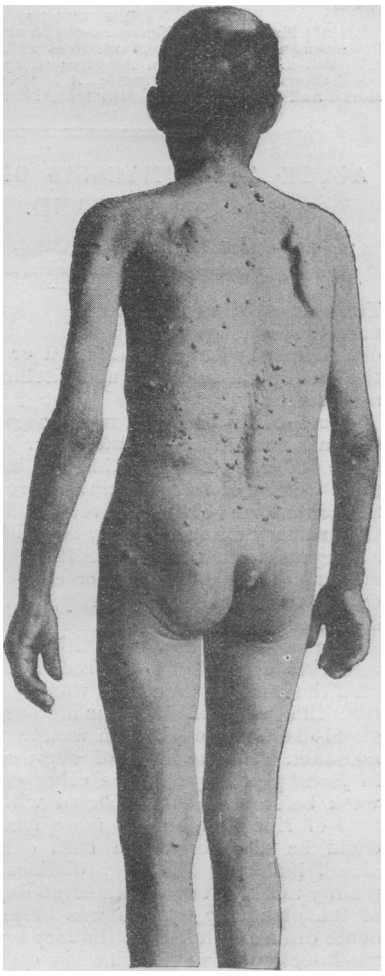
On several occasions he has pricked them, thinking they were little bladders with fluid contents. One continued to grow from the region of the lower costal margin on the right side until it reached almost to Poupart's ligament. This tumour was removed at the North Staffordshire Infirmary twenty-two years ago. It weighed 5¾ lb. Following on the operation, he states that the tumours increased rapidly in size and number, especially on the chest.

Although the disease was painless at the onset, he now states that he can tell for several days beforehand where fresh growths will occur, by a sensation of intense prickling suggesting "the bite of a gnat" which causes him to rub the part. There then appears a small violet-blue spot under the skin, which gradually becomes a small growth about the size of a small pin's head; it steadily increases in size or may remain stationary. On the scalp there is a large oval soft swelling measuring 5 in. by 3½ in. covered with a few hairs, and suggesting a bald patch in the accompanying photograph. This tumour, he has been informed, was present at the time of birth. Apart from bronchitis, his only complaint is cramp in the right leg after walking.

Distribution of the Growths.

The greater number of the growths are found on the trunk itself and especially on the back and chest. On the top of the head is the large soft tumour already mentioned. The face and forehead are dotted over with numerous small growths and there are innumerable small pin-point growths on the neck. There is one on the chin the size of a hazel nut, pedunculated, but hidden by the beard. The arms, thighs, and legs are comparatively free from growths. There are none on the palms of the hands, but on the sole of the left foot there are two small ones along the inner border, and several small ones on the sole of the right foot. Over the coccyx there is a tail-like appendage. The majority are sessile, but a few have a tendency to be pedunculated.

There is no pain on pressing over any of the tumours. They are generally of soft consistence, some giving the sensation of empty bags of skin, but over the site of the operation scar they tend to be firmer and give a feeling of containing a convoluted plexus. In most of the larger tumours this plexiform arrangement is distinctly palpable.



Areas of pigmentation are usually found associated with the tumour growth. In Alexis Thomson's series of 76 cases, pigmentation was present in 19—a proportion of 25 per cent. Harbitz[3] found pigmentation present in all the cases examined by him. In many cases, as in this, the skin surface generally is dark with a diffuse brownish pigmentation. In addition to this general discoloration, more definite areas of pigmentation are to be found. Three distinct varieties of pigmentation occur and are present in this case similar to those described by Rolleston and Macnaughton:

1. Punctiform pigment spots, resembling freckles.
2. *Café-au-lait* patches.
3. Violet-blue spots—the first stage of the molluscous tumours.

In this man the *café-au-lait* patches are found only on the back and especially over the scapulae. The largest is seen over the right scapula. Smaller patches occur on the back generally and over the buttocks. The arms and legs are comparatively free from pigmentation, and there are no patches on the buccal mucous membrane.

The actual diagnosis of von Recklinghausen's disease must, of course, rest on the histological examination of the tumours. This has not been agreed to in this case, and owing to the rapid increase which took place after the operation twenty-two years ago, I have not pressed him very hard to submit to any operation, however slight.

I have been quite unable to trace any hereditary or family tendency in this patient, but it is well known that such cases are not infrequent, and are fully described in the literature. As Feindel has said, "Generalized neurofibromatosis is always congenital, often hereditary, and sometimes familial."

The treatment for the condition is purely surgical, but should only be adopted in the case of very large tumours, if there is pain, or where ulceration has occurred over the tumour. It has repeatedly been observed that operation has been followed by an increase in the size and number of the tumours.[4]

Death may occur from exhaustion, loss of weight, and anaemia, if the disease is progressive, and the occurrence of sarcoma is not uncommon.

The special points of interest in this case are:

1. The intense itching preceding the appearance of the tumours. This symptom is not a common one, but was noted by Malcolm Morris and Wilfred Fox[5] in a case described by them.

2. The congenital nature of the disease, as shown by the presence of the tumour on the scalp at the time of birth.

3. The presence of tumours on the soles of the feet—an uncommon situation.
4. The presence of the three types of pigmentation.
5. The rapid increase in the size and number of tumours after operation.

REFERENCES
[1] Alexis Thomson: *On Neuroma and Neurofibromatosis*, 1900 [2] J. D. Rolleston and N. S. Macnaughton, *Review of Neurology and Psychiatry*, 1912, vol. x. [3] F. Harbitz, *Archives of Internal Medicine*, 1909, iii. [4] C. C. Choyce and J. M. Beattie: *A System of Surgery*, vol. iii. [5] M. Morris and W. Fox, *British Journal of Dermatology*, 1907, vol. xix.

ACUTE ACTINOMYCOSIS OF THE PAROTID GLAND.

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THE course of events after infection by the ray fungus is usually slow; the swelling and induration are of gradual development without acute local or general reaction, and the subsequent softening and discharge are long drawn out and tedious.

There occur, however, rarer cases in which the disease assumes a much more acute form. Two such cases have come under my notice within the last ten years, and they seem worthy of mention since there are but few reports of the acute forms of actinomycosis. In view of the fact that the ordinary chronic variety is still frequently missed, and a diagnosis, often of tuberculosis, sometimes of sarcoma, is wrongly made, the acuter cases of actinomycosis are, owing to their greater rarity, still more likely to be a source of error in diagnosis.

The two cases on which this note is based presented a remarkable similarity in onset and symptoms. In each case the route of infection was by the parotid duct, and from this point of entry the disease rapidly infiltrated the gland itself. The infection was, in the one case, derived, no doubt, from the habit of chewing corn whilst engaged in feeding poultry; in the other case the patient had, a week before the onset, played with some children in a field of ripe corn, but no more exact source of infection could be discovered. In both cases the outline of the gland, including the very distinct socia parotidis, was plainly marked, but within seven days of onset the barrier of the glandular capsule was broken down, and a most acute diffuse cellulitis of the face resulted. The nature of the infection was determined in each case by the examination of portions of the infiltrated subcutaneous tissue in the Pathological Department of the University of Manchester.

The features of this acute form of actinomycosis would appear to be as follows. The fungus enters by the parotid duct and, within a few days of entry, gives rise to an acute parotitis; the socia parotidis is seen to be enlarged and tender. The disease then bursts through the limits of the gland, whereupon a very acute cellulitis develops which may extend far over the scalp and well down the neck. There is great constitutional disturbance and marked evidence of septic absorption. At this stage the swollen parts are likely to be incised, when the incisions will be found to yield no pus, but merely a sanious debris. The cut tissue will be seen to be diffusely infiltrated, of a dirty grey colour, flecked with points of yellow. The fluid and debris obtained may be extremely foul, indicating a mixed infection of organisms from the mouth. The incisions, although they may ameliorate the acute condition, will probably fail to arrest the disease and the further spread of the lesion with fresh points of softening will be seen. These, when in turn incised, will tend to assume a chronic course until, ultimately, the diagnosis is thrust upon the observer by the yellow granules of the fungus.

The practical lesson is that any acute cellulitis of the face of obscure origin, or of unwonted appearance on incision, should excite suspicion of actinomycotic infection. The discharge from such lesions, or, better, a small portion of tissue, should be examined, and, since the fungus is not always easy of identification, a single negative result should by no means be accepted as final.

Once the diagnosis is established, appropriate treatment in addition to the incisions should be given. Iodine has appeared to yield good results in the more common chronic forms of the disease, and should certainly be used freely in these more acute cases. It has often been given in too small a dose; it should be pushed very freely in all forms of actinomycosis. One of my acute cases took no less than 240 grains of potassium iodide each day for several weeks with distinct benefit and no ill effect. In addition, iodine should be given locally. A 10 per cent. solution of iodipin may be injected into the infiltrated area at several points to the daily amount of 10 c.cm. The incisions and sinuses should be irrigated freely with a weak mixture of tincture of iodine and water.

In my second case a dose of 0.3 gram neo-salvarsan was given as soon as the diagnosis was established on the twelfth day of the illness. Although this case was a very acute one, with extremely foul discharge, there was within twenty-four hours of the administration a very marked increase in the amount of discharge and a rapid improvement in the local and general condition. This improvement was so well maintained that a further injection which was contemplated was not given. An isolated observation of this kind is of small value, but from my experience of its effect in this instance I would certainly be disposed to use salvarsan in cases of actinomycosis.

The problem of securing good drainage by incision of the face without leaving very obvious disfigurement is not easy. In my second case I attempted it by making a free incision above the hair line of the temple, and by blunt subcutaneous dissection I made a large tunnel, out of which a tube was drawn through a small vertical incision in front of the ear. Another incision, concealed behind the angle of the jaw, enabled a similar subcutaneous tunnel to reach the first one and provided good dependent drainage by a second tube. In this patient all the parts were soundly healed within two months and the cosmetic result was particularly fortunate.

In these more acute instances of facial actinomycosis early diagnosis is much to be desired. Unless the true nature of the disease is recognized, the condition is likely, after numerous incisions, to drift into the ordinary chronic form with multiple and tedious points of softening causing gross disfigurement. It would seem that much may be done to obviate this unfortunate result by energetic treatment early in the disease.

My first case had been treated as "mumps" and had been in existence for three weeks when brought to my notice. This case, in spite of energetic treatment, took five months to heal, and resulted in very marked disfigurement. The second case, which had a history of only seven days, was, by the help of free drainage, of much iodine, and probably of salvarsan, well healed within eight weeks, and the subsequent facial appearance of the patient was not the least satisfactory feature of the case.

Memoranda:

MEDICAL, SURGICAL, OBSTETRICAL.

A CASE OF ACETYL SALICYLIC ACID POISONING.

ACETYL SALICYLIC ACID being so extensively prescribed at the present time leads me to publish the following as possessing some features of interest in that the patient showed a marked idiosyncrasy to the drug on a number of occasions.

A gentleman, aged 34, was suffering from a slight attack of influenza. I advised him to take 10 grains of acetyl salicylic acid every six hours. The first dose was taken about half an hour after a light lunch, and by mistake he took only 5 grains. Within an hour his throat commenced to swell, and the mucous membrane of the tongue shortly afterwards became involved to a marked degree. Severe pain, situated over the middle of the sternum, ensued. The oedema spread to the neck, which became much enlarged equally on the two sides, and the swelling quickly extended upwards over the face. The eyelids participated in the general oedema, but the involvement was not sufficient to close the palpebral fissures. A dull red urticarial rash now appeared over the chin and both cheeks. The roof of the mouth was dry and the speech thick. The oedema of the tongue was sufficient to embarrass somewhat respiration by the mouth, but the act was unimpeded by the nasal route. Slight deafness and tinnitus and a sensation of fullness in the head were present. There was no palpitation, and the urine was normal in appearance. Gastro-intestinal symptoms, with the exception of the sternal pain mentioned, were absent. Two hours after the onset the symptoms commenced to abate, and an hour later had all disappeared. Although the discomfort